Case reports


Malignant gonadal tumour formation in intersexual states

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Summary
Two cases of malignant tumour are reported in phenotypically male hermaphrodites. The importance of establishing the presence of persistent Müllerian duct structures in pseudo-hermaphrodites is discussed in relation to prophylactic castration in anticipation of malignant change.

Introduction
This paper describes two cases of tumour formation in the gonads of hermaphrodites, which is a rare complication of an uncommon condition. The first patient went unrecognized as an intersexual state for many years. The second case is of a malignant ovarian tumour with normal fallopian tube in the scrotum of an outwardly normal male, and may be unique.

Case 1
M.M. was born with a moderately severe hypospadias and an absent right testicle. He was raised as a boy, and when 5 years old he started a series of staged urethroplasty operations. In addition, a short course of male hormone was given in an attempt to produce testicular descent on the right side. At the age of 14 a mammoplasty was performed for rightsided gynaecomastia. At the age of 16 his mother reported a brief episode of haematuria, and at this time his sexual status was questioned.

In view of the patient's physique, hair distribution and ability to achieve erection, he was judged to be a male. At the age of 21 the patient received psychiatric help for an anxiety neurosis, which continued for the rest of his life. At the age of 27 he attended the surgical outpatients' department complaining of a painful swelling in the left testicle. Physical examination revealed an absent right testicle; the left testicle was replaced by a slightly tender indurated mass 8 cm in diameter. The penis was small and was scarred by previous urethroplasty. In addition, the patient was short and spoke with a high pitched voice. Exploration of the scrotum showed a haemorrhagic tumour and an orchidectomy was performed. There was no clinical evidence of distant metastases, and lymphangiography showed no abnormality of the abdominal nodes. Histological examination showed a malignant teratoma (see Fig. 1).

Postoperatively a course of radiotherapy was given to the penis and para-aortic glands (4300 rad). Six months later a scrotal recurrence was treated by an iridium implant (4600 rad) with regression. Fifteen months following the original operation, pulmonary
metastases developed and the scrotal recurrence reappeared as a fungating mass. A further course of external irradiation (7000 rad) was unsuccessful. Accordingly, chemotherapy was undertaken at monthly intervals. Each course consisted of 5 fluorouracil 420 mg/day for 5 days; cyclophosphamide 420 mg, vincristine 1·4 mg, methotrexate 42 mg on days 1 and 4.

Initially there was a reduction in the size of the scrotal recurrence, but after 4 months the tumour metastases again increased in size. The patient died 30 months after the original operation.

At post-mortem examination there were extensive pulmonary metastases and local invasion around the scrotal recurrence. A rudimentary uterus and cervix were present. The precise opening of the cervix was destroyed by tumour. Neither a right-sided testicle nor any ovary was found.

Case 2

J.S., a 71-year-old man of short stature and with slight gynaecomastia, presented with a long standing hydrocoele. Physical examination showed otherwise normal external genitalia. Following aspiration of the scrotal swelling a painless irregular mass remained measuring 8 cm × 5 cm. The patient refused a definitive operation, and, therefore, regular aspirations were performed over a period of 13 years. Eventually permission for operation was given. A cystic mass 10 cm in diameter was removed. On the upper aspect of the mass was a vestigial fallopian tube. Histological examination of the cyst revealed a pseudomucinous cystadenocarcinoma of the ovary (Fig. 2). There was no testicular tissue present in this gonad, but exploration of the other side showed a normal testis and vas deferens. A biopsy of the testicular body revealed normal spermatogenesis to be present.

Buccal mucosa smears showed a chromatin negative pattern. The patient failed to attend for follow-up examination, and died 6 months later from a cerebral haemorrhage. Permission for a post-mortem examination was refused.
Discussion

Sporadic reports occur of phenotypically male subjects with persistent Müllerian duct structures. The first patient showed many clinical features of pseudo-hermaphroditism. The short stature, gynae- mastia, hypospadias, absent right testicle, and psychosexual disturbance are typical of the syndrome. The development of a post-pubertal teratoma might have been a chance occurrence developing in a normally positioned testicle, but it is probable that this was a dysgenetic testicle since it lacked the ability to produce a Müllerian duct-inhibiting factor.

Organ culture of the reproductive tract of the castrated rat has shown a Müllerian duct-inhibiting factor to be produced by the normal human fetal testis. This factor is distinct from androgen (Josso, 1972). The reports of cases in siblings have suggested the possibility of a genetically inherited absence of this factor (Brook et al., 1973). Another syndrome is described in which persistent Müllerian structures are associated with additional external genital malformation, and either total absence of one gonad or a 'residual streak'; this has been termed 'mixed gonadal dysgenesis' (Salle and Hedinger, 1970). This separation into two different syndromes is possibly artificial (Alexander and Ferguson Smith, 1961).

Tumour formation is well recognized in the mal-descended testicle or residual streak of patients with persistent Müllerian structures (Taub, 1954; Cornet, 1971). Whether the persistence of Müllerian structures increases the risk over that accepted for conventional cryptorchial patients or not is open to question (Brook et al., 1973; Cornet, 1971). Some writers feel the risk is sufficiently increased to justify prophylactic castration (Jirasek, 1971). Others have gone so far as to suggest conversion to a female phenotype as well as castration (Salle and Hedinger, 1970).

Cases of apparently simple hypospadias should be subject to buccal smear chromat in screening. Where other abnormalities of the external genitalia exist, more detailed chromosome studies should be undertaken. Where there is continued doubt into the nature of an intersex state, it would seem reasonable to establish the presence of persistent Müllerian duct structures by laparotomy or laparoscopy. The first case suggests the wisdom of castration before puberty. However, conversion to a female phenotype seems unjustifiable in view of the likely failure of psychological adjustment in the older patient (Dewhurst and Gordon, 1969).

The second case is most unusual. True hermaphroditism or gonadal intersexuality is rare, and requires the demonstration of both ovarian and testicular tissue. A review of previous documented cases shows the usual arrangement to be a bilateral ovo-

testis (Overzier, 1963). However, an ovary with a contralateral testis does occur.

The usual external genital arrangement is a phallus with a penile urethra or hypospadias (Butler et al., 1969), but the complete male form does exist (Overzier, 1963). A presentation at 80 years of age is most unusual, but has been described (Roberts and Khajari, 1964). Sasaki and Makino (quoted by Butler) described the presence of a scrotal ovary and contralateral testis. True hermaphroditic sex chromatin patterns in the literature have been 53% (46 XY) and 12% (XY), with the remainder showing various degrees of mosaicism.

Cystadenocarcinoma of a scrotal ovary in a phenotypically male subject with otherwise normal external genitalia does not appear to have been reported before. An example of a theca cell tumour of the ovary has, however, been described (Vaughn and Gonzales-Angulo, 1961). In intersex states with female phenotype, tumours of testicular origin have been described, but no increased incidence of ovarian tumours was shown (Fathalla, Nabil Raskad and Kerr, 1966).

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References


An unusual case of spontaneous bacterial myositis

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Summary
A further report of spontaneous bacterial myositis occurring in the British Isles is recorded. The case is unusual because both calf muscle compartments were simultaneously affected, and unique in that surgical decompression was required to prevent impending limb ischaemia.

Bacterial myositis rarely occurs among the indigenous population of temperate countries. In an account from Boston all three patients had recently arrived from tropical countries (Levin, Gardner and Waldvogel, 1971). Nevertheless, four fatal cases were seen in the post-mortem room of Addenbrookes Hospital in 1951–56 (Barrett and Gresham, 1958), a further case was recorded in California in 1964 (Altrocchi, 1971) and more recently its occurrence in a Londoner of Irish extraction has been reported (Rogers, 1973).

In contrast bacterial myositis accounts for 3–4% of all surgical admissions in certain tropical areas (Horn and Masters, 1968).

The following case is considered worth reporting because of certain unusual features which may shed further light on the aetiology of this condition.

Case report
A 30-year-old unmarried mother of two children was admitted to hospital as an emergency. Four hours before admission she suddenly developed pain behind her left ankle which rapidly spread to the calf muscles of that leg. A few minutes later the right ankle became painful, followed by radiation of pain to the right calf. The pain rapidly became severe and she noted increasing swelling in both calves. There was no preceding history of trauma or of self-injection, although she occasionally probed with a needle small septic spots on her legs and arms. Two days before admission she had danced barefoot on a local beach known to be contaminated with sewage. She suffered from epilepsy but was erratic in taking her medication. She had, on three occasions, been admitted to hospital with drug overdosage. She was not taking the contraceptive pill.

The patient was an unkempt and poorly nourished woman, temperature 37°C, pulse 84 beats/min. Both calves were hot, tender and swollen. There were small bruises around both knee joints, a small pustule was present on the medial aspect of the right knee and numerous petechial haemorrhages over the dorsum of both feet (Fig. 1). The lower limb pulses were readily palpable and there was no venous engorgement. Both knee joints were held rigidly in <30° of flexion and movement at either knee or ankle joint produced severe calf pain. No effusion could be detected in either knee joint and the regional nodes were not enlarged. There was hypoaesthesia in both legs, from the mid-calf level which was more marked distally.

Investigations at this time: haemoglobin, 12·4 g/100 ml; white cell count, 8900/mm³ with a normal differential; ESR, 19 mm in the first hour (Westergren); an ECG and chest X-ray showed no abnormality. A provisional diagnosis of bilateral calf vein thrombosis was made.

Over the next few hours, signs of arterial insufficiency began to appear. Skin pallor, venous guttering and anaesthesia developed in both feet, the calf swelling increased and became more tense but there was no skin oedema. Aspiration of the knee joint did not produce any fluid and arthrograms excluded the possibility of a ruptured Baker's cyst. It became
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